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An atypical site of a retroperitoneal epidermoid cyst in a middle-aged woman

Mitura Kryspin[a,*], Romanczuk Mikolaj[a], Mitura Alicja[b]

[a] Department of General Surgery, Siedlce Hospital, Starowiejska 15, 08-110 Siedlce, Poland
[b] Department of Pediatrics, Mazovia Hospital in Siedlce, Poniatowskiego 25, 08-110 Siedlce, Poland

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ABSTRACT

INTRODUCTION: Retroperitoneal cysts are rare, usually asymptomatic, abdominal lesions. Epidermoid cysts developing in this space usually occur in middle-aged women and are incidentally discovered in the presacral region during ultrasound examination. Occasionally, cysts may arise from splenic tissue or adrenal glands and develop above the presacral area.
PRESENTATION OF CASE: We present the unusual location of a cyst in the retroperitoneal space in a 41-year-old woman admitted to hospital due to detection of a lesion in ultrasound imaging. A CT scan confirmed large intra-abdominal cyst. At operation, a large retroperitoneal thin-walled cyst with no evident arising point was discovered. Histologic analysis revealed epidermoid cyst.
DISCUSSION: Our patient presented with giant retroperitoneal cyst extending from the subhepatic region to the uterine and bladder. To our knowledge, this unusual location in adult has not been previously reported in the literature. In our case the lesion was adjacent to inferior vena cava and mesenteric vessel which required special attention during preparation and was technically demanding.
CONCLUSION: Surgery is the gold standard for the diagnosis and treatment of retroperitoneal epidermoid cysts. Successful treatment of benign retroperitoneal epidermoid cysts depends on appropriate diagnosis, careful operative technique, and adequate management of the underlying pathology.

1. Introduction

Mesenteric and retroperitoneal cysts are rare, usually asymptomatic abdominal lesions. These malformations can be located in every region of the retroperitoneum. Cysts of the retroperitoneal space ought to be distinguished from omental, mesenteric, splenic and enteric cysts despite their similar symptomatology.[1] Epidermoid cysts developing in the retroperitoneal space are quite rare and usually occur in middle-aged women.[2] Abnormal masses in the abdomen are often incidentally discovered during ultrasound examination. Most patients with retroperitoneal cysts are asymptomatic. However, some patients may present with unspecific symptoms resulting from a local mass effect (e.g. lower abdominal pain, constipation).[2]

Diagnostic imaging is helpful in making the diagnosis, although histopathologic examination is crucial in the final analysis of the lesion.[1] Therefore, surgical resection is indicated to establish a diagnosis and prevent eventual complications, such as hemorrhage, infection or rupture.[3]

2. Presentation of case

A 41-year-old woman with no previous medical history was admitted to hospital due to the detection of an abnormal mass in the abdomen discovered in ultrasound imaging. A well-circumscribed hyperechogenic tumor measuring approximately 117 mm × 69 mm with microcalcifications was found in the right iliac fossa. The patient complained of recurrent unspecific lower abdomen pain without any additional symptoms. Physical examination revealed a large, soft, palpable abdominal mass without tenderness, located in the right lower quadrant of the abdomen. Laboratory tests revealed no abnormalities. A computed tomography (CT) scan showed a 171 mm × 113 mm × 93 mm cystic mass filled with homogeneous material (25 Hounsfield units; HU), with smooth boundaries and no contrast medium enhancement (Figs. 1 and 2) The lesion was located in front of the pancreas' head with inferior vena cava (IVC) compression, caudally extending to the urinary bladder and uterus. A mesenteric cyst was suspected. The patient underwent laparotomy. At operation, a large retroperitoneal thin-walled cyst with no evident arising point was discovered. We performed surgery with complete resection of the tumor mass and with macroscopic free margins. The cyst was excised after mobilization of the right ureter and mesenteric vessels, which were found on the anterior surface of the tumor. At surgery, special attention was paid to the relation between the lesion and the adjacent IVC. No enlarged lymph nodes were found. The cyst's dimensions were approximately 18 cm × 12 cm. In

* Corresponding author at: ul. Starowiejska 15, 08-110 Siedlce, Poland. Tel.: +48 602 809035.
E-mail address: chirurgia.siedlce@gmail.com (K. Mitura).

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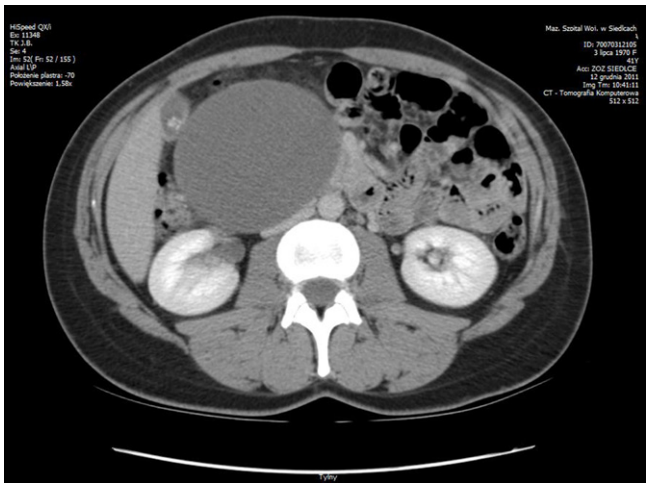

Fig. 1. Contrast-enhanced venous phase CT scan presenting an oval tumor arising in the subhepatic region.

histologic analysis, an epidermoid cyst was diagnosed with components of stratified squamous epithelium with a mix of desquamated material, cholesterol, and filled with keratinized cellular debris. (Figs. 3 and 4) The patient was discharged from hospital four days later, after an uneventful postoperative period.

3. Discussion

Retroperitoneal cysts which develop within the retroperitoneal space, externally from essential organs within that region, are rare intra-abdominal tumors with an incidence of 1 per 140 000 in surgery departments, and 1 per 20 000 in pediatric departments.[4] These tumors originate from an embryologic error during development.[5] Extensive use of computed tomography for diagnosis of abdominal and retroperitoneal malformations has enhanced the detection rate for these lesions. Among the different types of cystic masses located in the retroperitoneal space non-neoplastic or neoplastic lesions may be found. The most common non-neoplastic lesion is the pancreatic pseudocyst, characterized by round or oval encapsulated collections of pancreatic fluid, usually found in the peripancreatic region. Non-pancreatic pseudocysts manifest in CT as unilocular or multilocular fluid-filled

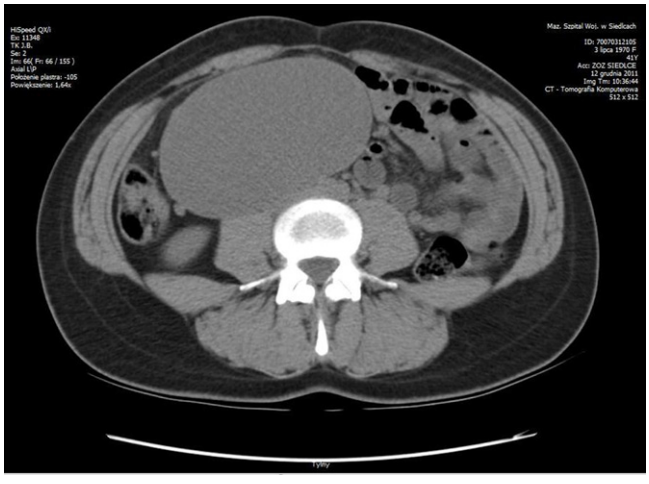

Fig. 2. Contrast-enhanced venous phase CT scan presenting a cyst filled with homogeneous material (25 HU).

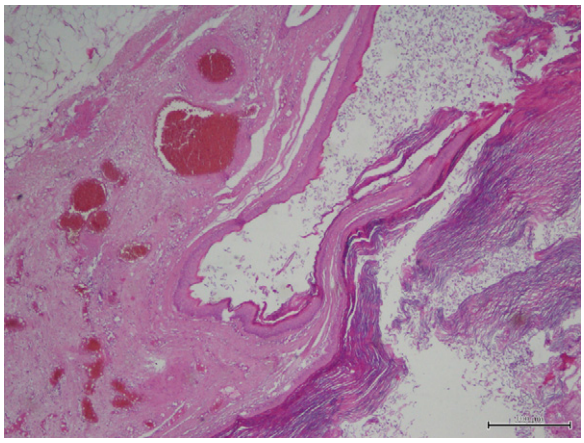

Fig. 3. Low-power histologic examination (100×) of an epidermoid cyst with components of stratified squamous epithelium with a mix of desquamated material.

masses with thick walls with no enzyme elevation.[6] Lymphocele, urinoma, hematoma and abscess are other non-neoplastic lesions. In differential diagnosis of these formations clinical history can be helpful in making an accurate diagnosis.[7] Neoplastic lesions include cystic lymphangioma, mucinous cystadenoma, cystic teratoma, or mesothelioma, müllerian cyst, epidermoid cyst, tailgut cyst, bronchogenic cyst or cystic change in solid neoplasms.[7] Many overlapping features have been shown to exist among the various retroperitoneal cystic masses. However, familiarity with the most significant radiologic characteristics, in correlation with clinical data, leads to correct lesion classification. An epidermoid cyst is a unilocular malformation, slow growing, and formed by desquamation of epithelial cells.[8] These lesion are commonly found in the presacral region in middle-age women.[9] An uncommon location of an epidermoid cyst outside of the presacral space was reported in an 11-year-old boy by Hagr in the subdiaphragmatic region.[10] Horn described an epidermoid cyst arising from an accessory spleen of the pancreas.[11] Benign extraadrenal epidermoid cysts mimicking adrenal tumors in abdominal imaging were reported by Grabellus.[12] Our patient presented with a giant retroperitoneal cyst extending from the subhepatic region to the uterus and bladder. To our knowledge, this unusual location in an adult has not been previously reported in the literature.

Ultrasound and computed tomography imaging did not allow us to unambiguously identify the lesion. Because it is impossible to entirely rule out cystic malignancies, surgical resection and histopathologic examination is required for definitive

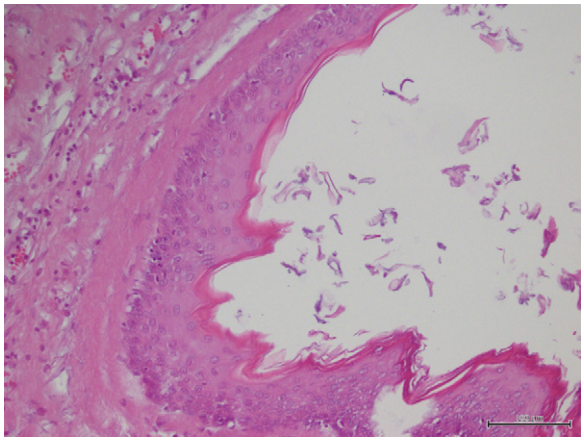

Fig. 4. High-power histologic examination (400×) presenting a cyst filled with keratinized cellular debris, surrounded by epithelium.

diagnosis. Therefore, explorative laparotomy was performed. Histologic examination revealed all of the main features of an epidermoid cyst. Consequently, an apparent diagnosis was made. The sole symptoms reported by the patient resulted from the compressive effect of the cyst on surrounding structures. No additional complications had developed before the operation, including inflammation, abscess or rupture. The incidence of recurrence for retroperitoneal cysts is higher than with other forms of cysts because their proximity to major blood vessels and other organs makes them difficult to completely excise.[13] In our case the lesion was adjacent to IVC and mesenteric vessels, which required special attention during preparation and was technically demanding.

4. Conclusion

Retroperitoneal location of an epidermoid cyst, outside the presacral region is extremely rare. Surgery is the gold standard for the diagnosis and treatment of retroperitoneal epidermoid cysts. Successful treatment of a benign retroperitoneal epidermoid cyst depends on appropriate diagnosis, careful operative technique and adequate management of the underlying pathology.

Conflict of interest

None.

Funding

None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

All authors contributed to study design and writing.

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